

*Questions
& Answers
about . . .*

Vitiligo

*National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)
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What Is Vitiligo?

Vitiligo (vit-ill-EYE-go) is a pigmentation disorder in which melanocytes (the cells that make pigment) in the skin, the mucous membranes (tissues that line the inside of the mouth and nose and genital and rectal areas), and the retina (inner layer of the eyeball) are destroyed. As a result, white patches of skin appear on different parts of the body. The hair that grows in areas affected by vitiligo usually turns white.

The cause of vitiligo is not known, but doctors and researchers have several different theories. One theory is that people develop antibodies that destroy the melanocytes in their own bodies. Another theory is that melanocytes destroy themselves. Finally, some people have reported that a single event such as sunburn or emotional distress triggered vitiligo; however, these events have not been scientifically proven to cause vitiligo.

Who Is Affected by Vitiligo?

About 1 to 2 percent of the world's population, or 40 to 50 million people, have vitiligo. In the United States, 2 to 5 million people have the disorder. Ninety-five percent of people who have vitiligo develop it before their 40th birthday. The disorder affects all races and both sexes equally.

Key Words

Antibodies—protective proteins produced by the body’s immune system to fight infectious agents (such as bacteria or viruses) or other “foreign” substances. Occasionally, antibodies develop that can attack a part of the body and cause an “autoimmune” disease. These antibodies are called autoantibodies.

Pigment—a coloring matter in the cells and tissues of the body.

Pigmentation—coloring of the skin, hair, mucous membranes, and retina of the eye.

Depigmentation—loss of color in the skin, hair, mucous membranes, or retina of the eye.

Melanin—a yellow, brown, or black pigment that determines skin color. Melanin also acts as a sunscreen and protects the skin from ultraviolet light.

Melanocytes—special skin cells that produce melanin.

Ultraviolet light A (UVA)—one type of radiation that is part of sunlight and reaches the earth’s surface. Exposure to UVA can cause the skin to tan. Ultraviolet light is also used in a treatment called phototherapy for certain skin conditions, including vitiligo.

Vitiligo seems to be more common in people with certain autoimmune diseases (diseases in which a person's immune system reacts against the body's own organs or tissues). These autoimmune diseases include hyperthyroidism (an overactive thyroid gland), adrenocortical insufficiency (the adrenal gland does not produce enough of the hormone called corticosteroid), alopecia areata (patches of baldness), and pernicious anemia (a low level of red blood cells caused by failure of the body to absorb vitamin B₁₂). Scientists do not know the reason for the association between vitiligo and these autoimmune diseases. However, most people with vitiligo have no other autoimmune disease.

Vitiligo may also be hereditary, that is, it can run in families. Children whose parents have the disorder are more likely to develop vitiligo. However, most children will not get vitiligo even if a parent has it, and most people with vitiligo do not have a family history of the disorder.

What Are the Symptoms of Vitiligo?

People who develop vitiligo usually first notice white patches (depigmentation) on their skin. These patches are more common in sun-exposed areas, including the hands, feet, arms, face, and lips. Other common areas for white patches to appear are the armpits and groin and around the mouth, eyes, nostrils, navel, and genitals.

Vitiligo generally appears in one of three patterns. In one pattern (focal pattern), the depigmentation is limited to one or only a few areas. Some people develop depigmented patches on only one side of their bodies (segmental pattern). But for most people who have vitiligo, depigmentation occurs on different parts of the body (generalized pattern). In addition to white patches on the skin, people with vitiligo may have premature graying of the scalp hair, eyelashes, eyebrows, and beard. People with dark skin may notice a loss of color inside their mouths.

Will the Depigmented Patches Spread?

There is no way to predict if vitiligo will spread. For some people, the depigmented patches do not spread. The disorder is usually progressive, however, and over time the white patches will spread to other areas of the body. For some people, vitiligo spreads slowly, over many years. For other people, spreading occurs rapidly. Some people have reported additional depigmentation following periods of physical or emotional stress.

How Is Vitiligo Diagnosed?

If a doctor suspects that a person has vitiligo, he or she usually begins by asking the person about his or her medical history. Important factors in a person's medical history are a family history of vitiligo; a rash, sunburn, or other skin trauma at the site of vitiligo 2 to 3 months before depigmentation started; stress or physical illness; and premature (before age 35) graying of the hair. In addition, the doctor will need to know whether the patient or anyone in the patient's family has had any autoimmune diseases and whether the patient is very sensitive to the sun. The doctor will then examine the patient to rule out other medical problems. The doctor may take a small sample (biopsy) of the affected skin. He or she may also take a blood sample to check the blood-cell count and thyroid function. For some patients, the doctor may recommend an eye examination to check for uveitis (inflammation of part of the eye). A blood test to look for the presence of antinuclear antibodies (a type of autoantibody) may also be done. This test helps determine if the patient has another autoimmune disease.

How Can People Cope With the Emotional and Psychological Aspects of Vitiligo?

The change in appearance caused by vitiligo can affect a person's emotional and psychological well-being and may create difficulty in getting or keeping a job. People with this disorder can experience emotional stress, particularly if vitiligo develops on visible areas of the body, such as the face, hands, arms, feet, or on the genitals. Adolescents, who are often particularly concerned about their appearance, can be devastated by widespread vitiligo. Some people who have vitiligo feel embarrassed, ashamed, depressed, or worried about how others will react.

Several strategies can help a person cope with vitiligo. First, it is important to find a doctor who is knowledgeable about vitiligo and takes the disorder seriously. The doctor should also be a good listener and be able to provide emotional support. Patients need to let their doctors know if they are feeling depressed because doctors and other mental health professionals can help people deal with depression. Patients should also learn as much as possible about the disorder and treatment choices so that they can participate in making important decisions about medical care.

Talking with other people who have vitiligo may also help a person cope. The National Vitiligo Foundation can provide information about vitiligo and refer people to local chapters that have support groups of patients, families, and physi-

cians (see page 17). Family and friends are another source of support.

Some people with vitiligo have found that cosmetics that cover the white patches improve their appearance and help them feel better about themselves. A person may need to experiment with several brands of concealing cosmetics before finding the product that works best.

What Treatment Options Are Available?

The goal of treating vitiligo is to restore the function of the skin and to improve the patient's appearance. Therapy for vitiligo takes a long time—it usually must be continued for 6 to 18 months. The choice of therapy depends on the number of white patches and how widespread they are and on the patient's preference for treatment. Each patient responds differently to therapy, and a particular treatment may not work for everyone. Current treatment options for vitiligo include medical, surgical, and adjunctive therapies (therapies that can be used along with surgical or medical treatments).

Medical Therapies

Topical Steroid Therapy

Steroids may be helpful in repigmenting the skin (returning the color to white patches), particularly if started early in the disease. Corticosteroids are a group

Treatment Options for Vitiligo

Medical Therapies

- Topical steroid therapy
- Topical psoralen photochemotherapy
- Oral psoralen photochemotherapy
- Depigmentation

Surgical Therapies

- Skin grafts from a person's own tissues (autologous)
- Skin grafts using blisters
- Micropigmentation (tattooing)
- Autologous melanocyte transplants

Adjunctive Therapies

- Sunscreens
- Cosmetics
- Counseling and support

of drugs similar to the hormones produced by the adrenal glands (such as cortisone). Doctors often prescribe a mild topical corticosteroid cream for children under 10 years old and a stronger one for adults. Patients must apply the cream to the white patches on their skin for at least 3 months before seeing any results. It is the simplest and safest treatment but not as effective as psoralen

photochemotherapy (see below). The doctor will closely monitor the patient for side effects such as skin shrinkage and skin striae (streaks or lines on the skin).

Psoralen Photochemotherapy

Psoralen photochemotherapy (psoralen and ultraviolet A therapy, or PUVA) is probably the most beneficial treatment for vitiligo available in the United States. The goal of PUVA therapy is to repigment the white patches. However, it is time-consuming and care must be taken to avoid side effects, which can sometimes be severe. Psoralens are drugs that contain chemicals that react with ultraviolet light to cause darkening of the skin. The treatment involves taking psoralen by mouth (orally) or applying it to the skin (topically). This is followed by carefully timed exposure to ultraviolet A (UVA) light from a special lamp or to sunlight. Patients usually receive treatments in their doctors' offices so they can be carefully watched for any side effects. Patients must minimize exposure to sunlight at other times.

Topical Psoralen Photochemotherapy

Topical psoralen photochemotherapy often is used for people with a small number of depigmented patches (affecting less than 20 percent of the body). It is also used for children 2 years old and older who have localized patches of vitiligo. Treatments are done in a doctor's office under artificial UVA light once or twice a week. The doctor or nurse applies a thin coat of psoralen to the patient's depigmented patches about 30

minutes before UVA light exposure. The patient is then exposed to an amount of UVA light that turns the affected area pink. The doctor usually increases the dose of UVA light slowly over many weeks. Eventually, the pink areas fade and a more normal skin color appears. After each treatment, the patient washes his or her skin with soap and water and applies a sunscreen before leaving the doctor's office.

There are two major potential side effects of topical PUVA therapy: (1) severe sunburn and blistering and (2) too much repigmentation or darkening of the treated patches or the normal skin surrounding the vitiligo (hyperpigmentation). Patients can minimize their chances of sunburn if they avoid exposure to direct sunlight after each treatment. Hyperpigmentation is usually a temporary problem and eventually disappears when treatment is stopped.

Oral Psoralen Photochemotherapy

Oral PUVA therapy is used for people with more extensive vitiligo (affecting greater than 20 percent of the body) or for people who do not respond to topical PUVA therapy. Oral psoralen is not recommended for children under 10 years of age because of an increased risk of damage to the eyes, such as cataracts. For oral PUVA therapy, the patient takes a prescribed dose of psoralen by mouth about 2 hours before exposure to artificial UVA light or sunlight. The doctor adjusts the dose of light until the skin areas being treated become pink. Treat-

ments are usually given two or three times a week, but never 2 days in a row.

For patients who cannot go to a PUVA facility, the doctor may prescribe psoralen to be used with natural sunlight exposure. The doctor will give the patient careful instructions on carrying out treatment at home and monitor the patient during scheduled checkups.

Known side effects of oral psoralen include sunburn, nausea and vomiting, itching, abnormal hair growth, and hyperpigmentation. Oral psoralen photochemotherapy may increase the risk of skin cancer. To avoid sunburn and reduce the risk of skin cancer, patients undergoing oral PUVA therapy should apply sunscreen and avoid direct sunlight for 24 to 48 hours after each treatment. Patients should also wear protective UVA sunglasses for 18 to 24 hours after each treatment to avoid eye damage, particularly cataracts.

Depigmentation

Depigmentation involves fading the rest of the skin on the body to match the already white areas. For people who have vitiligo on more than 50 percent of their bodies, depigmentation may be the best treatment option. Patients apply the drug monobenzyether of hydroquinone (monobenzone or Benoquin*) twice a day to

* Brand names included in this booklet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

pigmented areas until they match the already depigmented areas. Patients must avoid direct skin-to-skin contact with other people for at least 2 hours after applying the drug.

The major side effect of depigmentation therapy is inflammation (redness and swelling) of the skin. Patients may experience itching, dry skin, or abnormal darkening of the membrane that covers the white of the eye. Depigmentation is permanent and cannot be reversed. In addition, a person who undergoes depigmentation will always be abnormally sensitive to sunlight.

Surgical Therapies

All surgical therapies must be viewed as experimental because their effectiveness and side effects remain to be fully defined.

Autologous Skin Grafts

In an autologous (use of a person's own tissues) skin graft, the doctor removes skin from one area of a patient's body and attaches it to another area. This type of skin grafting is sometimes used for patients with small patches of vitiligo. The doctor removes sections of the normal, pigmented skin (donor sites) and places them on the depigmented areas (recipient sites). There are several possible complications of autologous skin grafting. Infections may occur at the donor or recipient sites.

The recipient and donor sites may develop scarring, a cobblestone appearance, or a spotty pigmentation, or may fail to repigment at all. Treatment with grafting takes time and is costly, and most people find it neither acceptable nor affordable.

Skin Grafts Using Blisters

In this procedure, the doctor creates blisters on the patient's pigmented skin by using heat, suction, or freezing cold. The tops of the blisters are then cut out and transplanted to a depigmented skin area. The risks of blister grafting include the development of a cobblestone appearance, scarring, and lack of repigmentation. However, there is less risk of scarring with this procedure than with other types of grafting.

Micropigmentation (Tattooing)

Tattooing implants pigment into the skin with a special surgical instrument. This procedure works best for the lip area, particularly in people with dark skin; however, it is difficult for the doctor to match perfectly the color of the skin of the surrounding area. Tattooing tends to fade over time. In addition, tattooing of the lips may lead to episodes of blister outbreaks caused by the herpes simplex virus.

Autologous Melanocyte Transplants

In this procedure, the doctor takes a sample of the patient's normal pigmented skin and places it in a labo-

ratory dish containing a special cell culture solution to grow melanocytes. When the melanocytes in the culture solution have multiplied, the doctor transplants them to the patient's depigmented skin patches. This procedure is currently experimental and is impractical for the routine care of people with vitiligo.

Additional Therapies

Sunscreens

People who have vitiligo, particularly those with fair skin, should use a sunscreen that provides protection from both the UVA and UVB forms of ultraviolet light. Sunscreen helps protect the skin from sunburn and long-term damage. Sunscreen also minimizes tanning, which makes the contrast between normal and depigmented skin less noticeable.

Cosmetics

Some patients with vitiligo cover depigmented patches with stains, makeup, or self-tanning lotions. These cosmetic products can be particularly effective for people whose vitiligo is limited to exposed areas of the body. Dermablend, Lydia O'Leary, Clinique, Fashion Flair, Vitadye, and Chromelin offer makeup or dyes that patients may find helpful for covering up depigmented patches.

Counseling and Support Groups

Many people with vitiligo find it helpful to get counseling from a mental health professional. People often find they can talk to their counselor about issues that are difficult to discuss with anyone else. A mental health counselor can also offer patients support and help in coping with vitiligo. In addition, it may be helpful to attend a vitiligo support group.

What Research Is Being Done on Vitiligo?

For more than a decade, research on how melanocytes play a role in vitiligo has greatly increased. This includes research on autologous melanocyte transplants. At the University of Colorado, NIAMS supports a large collaborative project involving families with vitiligo in the United States and the United Kingdom. To date, over 2,400 patients are involved. It is hoped that genetic analysis of these families will uncover the location—and possibly the specific gene or genes—conferring susceptibility to the disease. Doctors and researchers continue to look for the causes of and new treatments for vitiligo.

Where Can People Get More Information About Vitiligo?

- **American Academy of Dermatology**
P.O. Box 4014
Schaumburg, IL 60168-4014
Phone: 847-330-0230 or
888-462-DERM (3376) (free of charge)
Fax: 847-330-0050
www.aad.org

The academy is the national organization for dermatology. It is dedicated to achieving the highest quality of dermatologic care for everyone. The academy produces patient information on vitiligo. It can also provide referrals to dermatologists.

- **National Vitiligo Foundation**
700 Olympic Plaza Circle, Suite 404
Tyler, TX 75701
Phone: 903–595–3713
Fax: 903–593–1545
E-mail: info@nvfi.org
www.nvfi.org

The foundation strives to locate, inform, and counsel vitiligo patients and their families; to increase public awareness and concern for the vitiligo patient; to broaden the concern for the patient within the medical community; and to encourage, promote, and fund increased scientific and clinical research on the cause, treatment, and ultimate cure.

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